

CONGENITAL ORBITAL TERATOMA

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ABSTRACT

Congenital orbital teratoma though rare is available in this environment. This is a case report of a baby with a protruding orbital mass in the left eye with all classical clinical features of teratoma. Though the histopathological report fell short of confirming the diagnosis the clinical features and outcome of management strongly suggest that the lesion is a teratoma.

Multidisciplinary approach to the management not only saved the life of the baby in question but also enhanced the outcome of treatment. Good and compliant follow up for six months was experienced. Cytological test is mandatory for any suspected cases of teratoma.

Key Words: Congenital, Orbit, Teratoma.

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INTRODUCTION

Teratomas are tumors containing representative cells from all three embryonic layers: ectoderm, mesoderm and endoderm¹. Orbital teratomas are very rare². Given the considerable variety of tissues that make up the orbit, orbital tumors constitute a heterogeneous array of lesions, and as such pose serious challenges to the ophthalmologist. The case in point, though the histopathological report fell short of confirming the diagnosis, the clinical features and other investigations strongly support the diagnosis of an orbital teratoma in a three day old Nigerian baby of the Ibo extraction.

CASE REPORT

Baby P.E. was delivered in Federal Medical Center, Jalingo three days prior to presentation. The Female baby was referred to National Eye Hospital, Kaduna on the second day of delivery because of the abnormal, fleshy protrusion from the left orbit. The mother rather than go to Kaduna chose to come to Guinness Eye Center, Onitsha because both parents hail from this part of the country. The baby was seen in the hospital on the fourth day of delivery.

The mother gave a history of a nine month pregnancy in which she was treated for Typhoid fever and malaria at seven month gestational period with Amoxycillin, Amodiaquine and Artesunate. The delivery was normal. The patient is the second child of the parents. The first is alive and healthy.

Examination showed a healthy and normal baby except for the oculo-orbital lesion. The left eye and adnexa showed a protruding, fleshy and tubular mass and a shrunken eyeball. The mass was non tender,

fluctuant and markedly mobile, measuring 5cm in length and 2cm in the widest diameter. It was continuous with shrunken eyeball with chemosed conjunctiva and keratinized cornea. There was mucopurulent discharge. The right eye was essentially normal (see fig 1).

The baby was admitted and the following investigations carried out: Conjunctival swab for microscopy, culture and sensitivity. Orbital x-ray with different views was done. The baby was commenced on topical chloramphenicol therapy. The x-ray showed a left orbital soft tissue mass but no orbital enlargement and the swab showed no growth. The baby was referred to the Neuro-surgeon and Pediatrician for necessary evaluation. A CT-Scan was done without contrast and 3mm/5mm/10mm slices were obtained to cover the orbit, skull base and brain. The findings included a soft tissue mass arising from the orbit. The left eye ball was extra orbital. There was no defect in the orbital roof or floor nor discernible continuity with the intracranial cavity. This made the diagnosis of encephalocele doubtful. Septum cavum pellucidum was noted.

The baby was kept in the neonatology unit/ward for 3 weeks. After 3 weeks the baby had examination under anaesthesia (EUA) and partial exenteration (see fig 2). The neurosurgical team was in attendance in the theatre during the EUA and surgery ready for action should the need arise. There was no detectable communication between the orbital mass and the brain. The surgery was uneventful. The excised tissue was sent for histopathological test. The baby was discharged after one week and was followed up for up to three months post-operatively. At three months post-operation there was no sign of recurrence of the tumor. The baby had a prosthesis inserted after 3 months (see fig 3). Six months post-

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operation baby is still healthy, left orbit clean and prosthesis in place. The first Histopathological report showed an orbito-ocular mass with disorganized eyeball. No cytological report was offered. The specimen and slide were sent to another pathologist for confirmation. The second histopathological report confirms that the orbito-ocular mass is a teratoma. The histopathological findings are attached. Herewith:

Figure 1: Oculo-orbital Tumour (Left Eye)



Figure 2: Tumour Excised with Partial Exenteration.



Figure 3: Prosthesis in the Left Socket.



PATHOLOGICAL FINDINGS:

MACROSCOPY: Specimen consist of a grayish white mass measuring 4.2x3.4x1.6cm. Cut section show grayish white surfaces with cystic openings and focal areas of haemorrhages and necrosis.

MICROSCOPY; Section of orbital mass show a benign soft tissue haphazardly arranged and containing more than one germ cell layer. Most of its part is covered by stratified squamous cell epithelium. Within this are numerous proliferating congested and dilated vascular channels with proliferating neural bundles and some neural bundles lined by pigmented choroidal epithelium. Also seen are abundant fibrous tissues scattered within the sampled areas with focal areas of chronic inflammatory cell infiltration. Areas of haemorrhages and haemorrhagic necrosis are also seen. This picture is in keeping with a congenital benign teratoma of the eye.

Figure 4:X40 Showing Numerous Vascular Channels, Proliferating Neural Bundles and Fibroblasts.

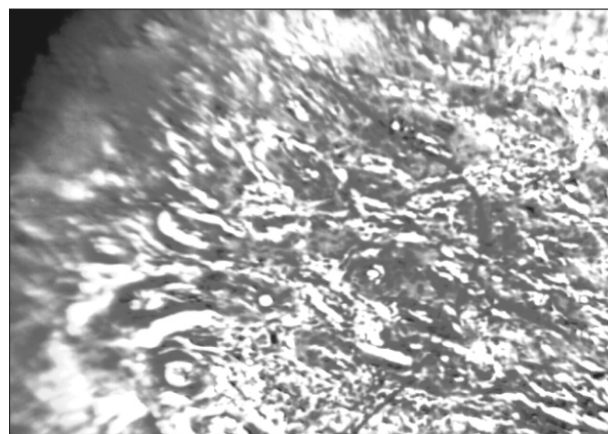


Figure 5: X100: Showing Stratified Squamous Epithelial cells towards the outside with Vascular Channels and Proliferating Fibroblasts.

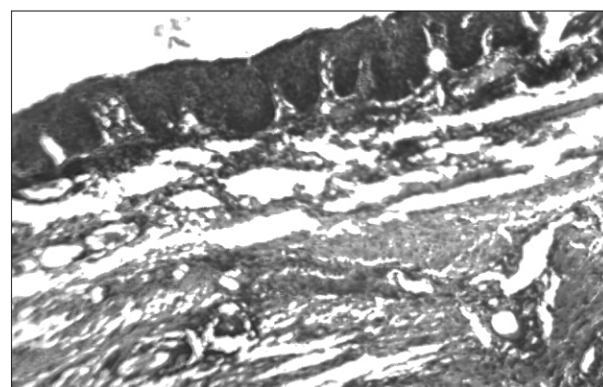


Figure 6: x100: Showing Neural bundles lined by Pigmented Choroidal Epithelium.

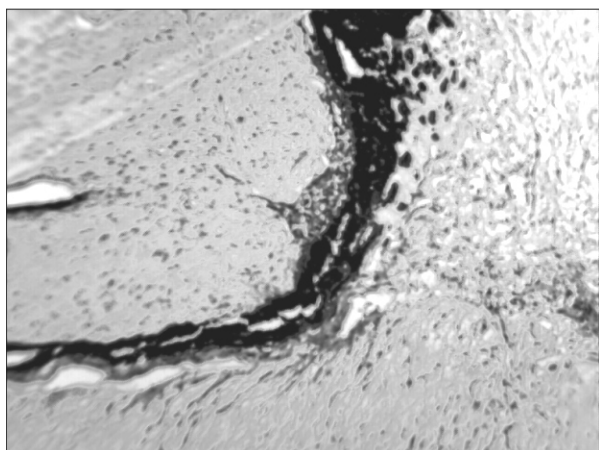
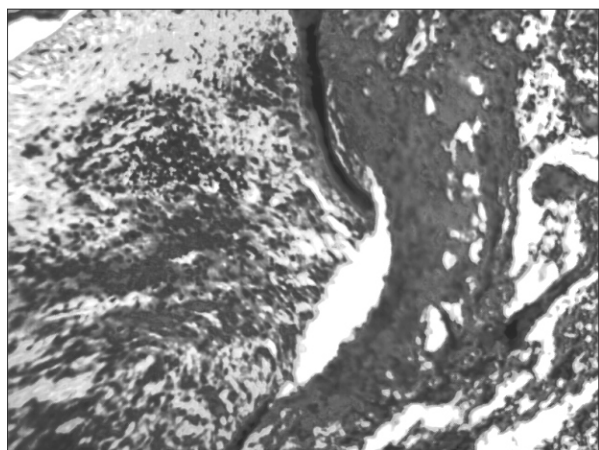


Figure 7: x100; Showing Proliferating Neural Bundles.



DISCUSSION

Though the first histopathological report failed to confirm the diagnosis of orbital teratoma in this report, the presentation at birth, the clinical features, unilaterality of the lesion, the radiological findings and the CT-Scan findings are all strongly in favour of orbital teratoma³. The rarity of the existence of orbital teratoma is not in doubt, previous studies have confirmed its presence in this environment^{4,5}. The failure to confirm the diagnosis in the first report by histopathological test is likely due to non inclusion of cytological test. Cytological test should be mandatory for any suspected case of teratoma. Proptosis or orbito-ocular protrusion at birth is not a common feature of orbital tumors⁶, so such rare tumors like teratoma must be ruled out once this occurs. The non-orbital enlargement seen in the x-ray in this case may be due to the fact that the tumor was already extraorbital.

The second histopathological report confirms that the orbito-ocular mass is a teratoma

Multi disciplinary management applied in this case

prevented misdiagnosis, enhanced good outcome of the treatment and saved the baby's life.

After six months post-operation there was no evidence of malignancy or recurrence. This is in keeping with congenital orbital teratoma which contains all germ cell layers and is usually benign and recurrent^{3,7}. Where clinical evidence of teratoma is overwhelming as in this case, management should be in line with the clinical diagnosis and unnecessary delay need to be avoided. Nevertheless cytological test should be done for any suspected case of teratoma.

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